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Lymphoproliferative malignancies in association with endemic African Kaposi's Sarcoma

ME STEIN, D SPENCER, P RUFF

SUMMARY

The association of classical Kaposi's sarcoma with lymphoproliferative disorders is well known. However, far less is known about lymphoproliferative malignancies in endemic African Kaposi's sarcoma. A review of 47 patients with the endemic type of Kaposi's sarcoma treated at the Johannesburg Teaching Hospital Complex between 1980 and 1992 revealed four patients (8,5 pc) in whom Kaposi's sarcoma was associated with a malignant lymphoma. Possible pathogenetic mechanisms are suggested and the current literature is reviewed.

Department of Medicine
Division of Clinical Haematology and Medical Oncology
University of Witwatersrand Medical School
7 York Road
Parktown 2193
Johannesburg, South Africa
Correspondence to:
P Ruff

INTRODUCTION

The close association between classical Kaposi's sarcoma (CKS) and lymphoproliferative malignancies (LPM) has been reported with a frequency of between 17 pc and 60 pc.[1] More recent studies have reported LPMs in up to one third of the patients with CKS.[2] Most lymphomas are of B-cell origin including chronic lymphocytic leukaemia, multiple myeloma, follicular and diffuse lympomas,[3] and appear to be associated with impaired immunological function.[4-6]

There were very few reports of the association between the endemic, African variant of Kaposi's sarcoma (AKS) and LPMs.[7] The scarcity of reports may, however, be due to socio-economic factors including lack of adequate medical facilities, poor compliance and follow up, as well as increased mortality and morbidity due to associated infectious events.

In this analysis, four patients with LPMs associated with AKS are analyzed.

MATERIALS AND METHODS

The records of all patients with AKS treated and followed at the Johannesburg Hospital Complex between 1980 and1992 were analyzed. On presentation the patients underwent physical examination, haematological and biochemical studies and chest radiography. When clinically indicated, further gastrointestinal and radiological investigations were performed. All patients were treated with either radiotherapaeutic or chemotherapeutic regimens as indicated.

RESULTS

Patient characteristics: Forty seven southern African (South Africa, Mozambican, Botswanan, Namibian and Malawian) patients were included in the study. All were male with a mean age of 53 years (range 24–82 years) and were HIV negative.

Associated lymphoproliferative malignancies: Three patients presented with generalised lymphadenopathy simultaneously with AKS. Lymph node biopsy revealed a malignant lymphoma in all three,

namely peripheral T-cell lymphoma, follicular lymphoma and immunoblastic lymphoma. In all three patients, the lymphoma was limited to the lymph nodes and responded to chemotherapy. No specific treatment was given for the AKS.

The fourth patient presented simultaneously with Kaposi's sarcoma involving peripheral lymph nodes, as well as hepatosplenomegaly, ascites and pleural effusion. Haematological and biochemical studies revealed a pancytopenia and pathological liver function tests. Immune function studies showed an absolute T-cell lymphopenia and a low T4/T8 ratio (0,78:1). Bone marrow trephine biopsy demonstrated infiltration with a B-cell lymphoma. Following cytarabine (cytosine arabinoside) and interferon-alpha therapy, a significant reduction in tumour bulk was achieved together with a marked symptomatic improvement. The patient was then continued on CHOP (cyclophosphamide, epirubicin, vincristine and prednisone) with further response, albeit of short duration.

The patient relapsed and died three months after the start of CHOP therapy with massive hepatosplenomegaly, pancytopenia and extensive lymphomatous infiltration of bone marrow.

DISCUSSION

Early reports from the 1960s suggested an association between AKS and lymphoproliferative malignancies. However, only a few sporadic cases have been reported.[8-10] One case of Hodgkin's disease was described among 19 autopsy cases of AKS.[10] Other authors[9,11-13] described sporadic cases of AKS associated with non-Hodgkin's lymphoma, Hodkin's disease or chronic lymphocytic leukaemia. Larger series, comprising hundreds of patients with AKS revealed very few cases of associated LPM.[7,8]

It is possible that the younger age of AKS patients, shorter survival due to accompanying infectious diseases and the lack of adequate medical facilities and follow up may be responsible for the lower frequency of LPM reported in AKS.[14-16]

The aetiology of AKS and associated lymphoproliferative disorders is not entirely clear. AKS is not associated with overt immunosuppression.[15] Recent reports, however, have demonstrated an alteration in cell mediated immunity and lymphocytic dysfunction in AKS.[18,19] In Africa, the coexistence of protein-calorie malnutrition as well as tropical and chronic infections may also result in impaired cellular immunity. T-cell regulatory dysfunction leading to unopposed proliferation of abnormal B-cells may also play a role.[19] In addition, the use of alkylating agents such as nitrogen mustard trenimon and cyclophosphamide in AKS may also cause immunosuppressive changes and result in the emergence of secondary lymphoproliferative malignancies.[20-22] This does not however, apply to our study as all four patients presented simultaneously with AKS and a malignt lymphoma, and did not receive specific treatment for their AKS.

In conclusion therefore, we have described four (8,5 pc) patients out of a total of 47 patients with AKS, who presented simultaneously with a LPM. Although no specific aetiology is known, abnormal T-cell and B-cell function in AKS, associated with poor nutrition and chronic infections may play a role. Improved outcome and prolonged survival among patients with AKS may result in increased awareness and more diagnoses of associated LPMs.

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